Case reports

Unusual echocardiographic appearance of intracardiac thrombi in a patient with endomyocardial fibrosis

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SUMMARY A five year old girl presented with a four month history of recurrent heart failure, which subsequently proved to be caused by endomyocardial fibrosis. There was no evidence of valvar disease. Echocardiography showed several echogenic masses with echolucent centres within the cardiac cavity. Histological examination showed that these masses were partly organised thrombus.

Endomyocardial fibrosis is rarely found in a patient who has never been to the tropics. The classic clinical course is that of a short lived pyrexial illness followed by a period of weeks or months of progressive deterioration of cardiac function leading to the development of heart failure. It is typically complicated by valvar dysfunction (usually mitral regurgitation) and occasionally by intracavity thrombus formation. In our patient there was no evidence of valvar dysfunction but there was progressive heart failure.

Case report

A five year old Greek girl presented with an acute exacerbation of a four month illness. This had begun with a two day episode of fever and dyspnoea that led to her being admitted to a local hospital where left ventricular failure was diagnosed and treated with digoxin and diuretics. Initially she had recovered well, but she was readmitted on several occasions with exacerbations of heart failure, with digoxin toxicity, or with chest infections.

She presented to this hospital because her illness had become worse over the preceding two weeks. She had become weak, sleepless, and had begun to vomit. She was anorexic and had some abdominal pain. Her parents noted that she had begun to look

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rather pale and yellow.

On examination she was obviously ill, slightly jaundiced, but apyrexial. Her blood pressure was 120/80 mm Hg and she had a tachycardia of 120 beats/min. The jugular venous pressure was raised and her liver, which was enlarged 7 cm below the costal margin, was smooth and soft. The spleen was not palpable. A third heart sound was heard but there were no signs of valvar heart disease.

The electrocardiogram showed a sinus tachycardia of 140 beats/min with some left axis deviation (-90°) . Voltage criteria indicated a left atrial abnormality and left ventricular hypertrophy. The chest x ray showed signs of left heart failure with upper lobe diversion of blood and pulmonary plethora, and there was appreciable cardiomegaly (cardiothoracic ratio 12.5:19 cm). Whole body x ray computerised tomographic scanning and ultrasound studies of the liver showed no evidence of a tumour.

A cross sectional echocardiogram was performed through the precordial, apical, and subcostal windows in standard projections. Both ventricles were dilated and globally hypokinetic. All valves appeared to be normal. At least five masses were seen within the left ventricle and one or more of them appeared to move freely about the ventricular cavities in a manner quite unlike that of a mural thrombus. Several appeared to be attached to the ventricular wall by a thin pedicle. There were at least two similar masses in the right ventricle, both of which did not appear to move within the cavity. The masses were spherical with echogenic perimeters; however, their

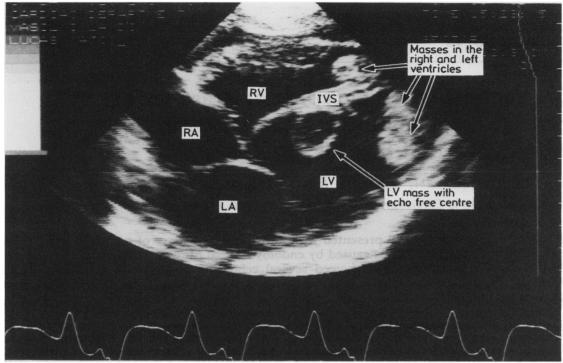


Figure A frame from the cross sectional echocardiogram via the subcostal window. This shows three of the masses with relatively echo-free centres lying within the cavity of the left ventricle and one mass at the apex of the right ventricle. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; IVS, interventricular septum.

centres were echo free. When the masses were viewed in real time the perimeters were seen to "wobble" in a way that suggested that these structures were easily deformed. The figure shows a frame recorded from the subcostal window.

In view of the haemodynamic risk of such a tumour mass in the ventricles and because of the need to establish a tissue diagnosis, the patient had left ventriculotomy with biopsy of both the myocardium and the intracardiac material. At operation the surgeon found that the interior of the left ventricle contained a large amount of recent blood clot, which was aspirated. There was frond-like material in the clot that was thought to be organised clot or possibly tumour tissue. The wall of the ventricle was found to be roughened and yellowish after aspiration of this material.

On the day after operation the patient deteriorated and died. Histological examination of the intracardiac material showed that it was an organising thrombus; there was no evidence of tumour. Vascular granulation tissue had filled up the intertrabecular spaces and the underlying myocardium showed some interstitial scarring but was otherwise unremarkable. Acute phase endomyocardial fibrosis was diagnosed at necropsy.

Discussion

In this patient the diagnosis of endomyocardial fibrosis was not suspected on clinical grounds. Although the clinical course was compatible with endomyocardial fibrosis, the fact that the patient had never been to the tropics made the diagnosis very unlikely. Echocardiography was undertaken as a part of the routine investigation of unexplained heart failure, and the finding of intracavity masses was unexpected. In view of the progressive ill health of the patient, the working diagnosis was that of tumour, either primary or secondary. Physical examination and whole body x ray computed tomography showed no other site of tumour, and, therefore, operation was undertaken both for diagnostic and therapeutic reasons.

Echocardiography is the best method for imaging intracardiac structures and for the detection of intracardiac masses. Cross sectional echocardiographs showing masses with relatively echo-free centres

have been described in patients with cystic tumours, but only a few of these have been caused by intracavity masses²⁻⁴—these were cases of atrial myxomas with cysts or areas of haemorrhage within them. There are no reports of appearances quite like those seen in our patient. Initially this echocardiograph was interpreted as showing multiple tumours with liquid, possibly necrotic, centres.

Review of the echocardiograph in the light of the findings at operation and necropsy indicated that the appearances were due to recently formed thrombus moving freely in the ventricle; the centre of the thrombus was not yet organised and was echo-free. We believe that the wobbly appearance of the perimeter was caused by a thin shell of organised thrombus surrounding a liquid centre.

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